Adenoid Cystic Carcinoma Presenting as Abducens Nerve Palsy Secondary to Cavernous Sinus Lesion

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Keywords

Adenoid cystic carcinoma · Cavernous sinus · Abducens nerve palsy

Abstract

Background: We report a 61-year-old who presented with a right-sided abducens nerve palsy secondary to a middle cranial fossa adenoid cystic carcinoma (ACC) extending into the cavernous sinus. **Purpose:** This case represents a unique presentation of intracranial ACC with a large middle cranial fossa mass and only a small extracranial component. **Methods:** Review of the literature was undertaken to identify cases of intracranial ACC and their range of presentations. **Results:** Our results show that this is the first reported case of an ACC presenting mostly as an intracranial mass with an isolated cranial nerve lesion. **Conclusion:** Our case highlights the importance of a broad differential diagnosis, particularly in circumstances where there are atypical features of lesions on radiographic imaging.

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Case Report

A 61-year-old female was referred to our hospital following a 3-week history of headaches and diplopia. She had no significant past medical history and was a non-smoker. Examination revealed right abducens nerve palsy. Neurological examination was otherwise unremarkable. Initial investigation with a contrast-enhanced CT revealed a homogeneously enhancing extra-axial mass in the medial aspect of the right middle cranial fossa involving the lateral wall of the cavernous sinus. There was no adjacent bone destruction or osteosclerosis.

Contrast-enhanced MRI demonstrated the homogeneously enhancing right middle cranial fossa mass filling the right cavernous sinus and extending medially into the pituitary fossa, as well as encasing the internal carotid artery (ICA). There was compression of the mesial temporal lobe and edema. A small extracranial component of the mass was noted, with extension through foramen ova-

Fig. 1. Pre-operative pre-contrast axial MRI T1-weighted (a) and T2-weighted (b) sequences showing a heterogeneous isointense right middle cranial fossa mass, with a small area of hyperintensity on the lateral aspect and a small amount of surrounding edema.

Fig. 2. Pre-operative post-contrast MRI T1 weighted sequencing in axial (a), sagittal (b), and coronal (c) views showing a homogeneously enhancing right middle cranial fossa mass involving the cavernous sinus.

le into the infratemporal fossa. The lesion was largely isointense on both T1- and T2-weighted imaging, with a small peripheral area of hyperintensity on T2-weighted sequences. The lesion was homogeneously contrast enhancing post administration of gadolinium (see Figures 1a, b and 2a–c).

The patient underwent a right temporal craniotomy limited debulking of the lesion via a sub-temporal approach. Histological examination of formalin-fixed tissue fragments showed that the tumor was composed of basa-

loid cells with a small amount of cytoplasm. Epithelial and myoepithelial cells were the 2 cells types evident in the sample, predominantly arranged cribriform structures and pseudocysts, lined by basement membrane material, and containing mucin. These histological findings were consistent with adenoid cystic carcinoma (ACC) (see Figure 3a–d).

Postoperatively she underwent contrast CT of neck, chest, abdomen, and pelvis, which did not show any evidence of a primary malignancy, specifically no lymph-

Fig. 3. Histological preparations of biopsy fragments with hematoxylin and eosin staining at 20× (a, c) and 40× (b, d) magnification. Both demonstrate the epithelial and myoepithelial cells arranged in a cribriform structure with pseudocysts, containing basement membrane.

adenopathy or enlarged salivary glands. A diagnosis of primary salivary gland ACC with peri-neural spread to the skull base and cavernous sinus was made.

Discussion

We present a patient with a cavernous sinus ACC secondary to peri-neural spread. This represents a rare cause of a middle cranial fossa extra-axial lesion and to our knowledge this is the first documented case of an ACC presenting as an intracranial mass with only a small extracranial component.

ACC is classified as a rare, malignant tumour arising from the salivary gland epithelium and accounts for approximately 10% of all epithelial salivary neoplasms. Together with mucoepidermoid carcinoma, ACC constitutes around one-half of all malignant salivary gland tumours [1].

Previously, it has been postulated that there are 3 main routes of intracranial spread of skull base ACCs: along the Eustachian tube (peritubal space), the mandibular and maxillary nerves, and the ICA [2]. This case would support this hypothesis with radiographical evidence of disease around and through the foramen ovale.

It has been documented that ACCs presenting with peri-neural spread carry a poorer prognosis than those identified as isolated lesions within the salivary glands [3]. However, overall survival is relatively poor, in the order of 10–20% at 15 years, due to the relentless nature of locoregional recurrence and the lack of efficacious treatment [4]. The rates of survival in patients with distant metastases are much lower, around 20% at 5 years [1].

There has been over 100 documented cases of ACC with intracranial involvement, with only a small portion of these having no evidence of a primary lesion [5]. Gormley et al. [6] reported a series of 16 patients with

ACC and intracranial extension, outlining the surgical management of these lesions, in particular relation to the ICA and the cavernous sinus. The illustrative case in this series regarding management of cavernous sinus ACC reports a case of a patient with a large nasopharyngeal mass extending through the skull base into the cavernous sinus. The nasopharyngeal mass was biopsied for diagnosis and then staged debulking, with an internal carotid vein graft, of the intracranial component was performed.

Similarly, Kyung et al. [7] presented a similar case of persistent abducens nerve palsy secondary to a nasopharyngeal mass extending into the cavernous sinus. This patient also underwent a biopsy of the nasopharyngeal component for diagnostic purposes.

Ginsberg and DeMonte [8] report 3 cases of palatal ACC with intracranial extension. One of these cases documents extension of an ACC into the cavernous sinus, from a hard palate mass invading into the pterygopalatine fossa and through the foramen rotundum.

It is likely in this case that the salivary gland malignancy has a primary extracranial lesion that has extended into the intracranial cavernous sinus. Another possibility is that of satellite cells that have seeded within the cavernous sinus through hematogenous spread, leading to a primary lesion within the cavernous sinus. Given the morphology of this particular lesion and its extension through the skull base into the infratemporal fossa, the former is more likely.

Our case represents a unique presentation of ACC with perineural spread to the cavernous sinus, in that there was no significant extracranial component amenable to biopsy prior to neurosurgical intervention.

Conclusion

ACC is a malignant tumor arising from salivary gland epithelium. We present a unique presentation of this disease, with a large intracranial and only a small extracranial component. Our case highlights the importance of a broad differential diagnosis, particularly in circumstances where there are atypical features of lesions on radiographic imaging.

Ethical Approval

Patient consent was obtained for the publication of this manuscript, in line with the institutional ethical protocols for case reports.

This article complies with ICMJE recommendations for conduct, reporting, editing, and publication of scholarly work in medical journals.

Author Contributions

Chris Donaldson and Bryden Dawes have done conception and literature review. Chris Donaldson has done drafting of article. Vivek Rathi has done collection and interpretation of histological images. Bryden Dawes has done review and final revisions. Kristian Bulluss has done final approval of the article.

Disclosure Statement

The authors declare that there are no conflicts of interest including financial, personal, or other relationships with people or organizations that would inappropriately influence their work. This research did not receive any specific grant from funding agencies in the public, private, or not-for-profit sectors.

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